EuropeanOphthalmicPathology Society
63rdAnnual Meeting
June 11 - 14, 2025
Basel, Switzerland
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Case number: 675/25
Materialdistributed: 1 histological slide

Meningioma in the Orbit

Clinicalhistory:17 yearsold boy wassent to theEyeClinicforgraduallyprogressingleftsidedproptosisconnectedwithparalyticstrabism and diplopia. His generalmedicalhistoryisunremarkable.Leftsidedproptosiswasapparentforthefirstlook, motility ofthelefteyewas very limited in thedirection to therightside, withsubsequentdiplopia. Theintraocularfindingsverequitenormal on botheyes, visualacuity 0,9 on therighteye, 0,7 on thelefteye. Intraocularpressurewas 20 mm/Hg on therightside, 21 mm/Hg on theleftside. Despiteofproptosis and problemswith motility thelefteye and entire orbital region lookedcalm, withoutinflammatorysigns, painless. Thefindings on theright orbit and righteyewasentirelynormal. MRI imagingwasindicated. It showedthe tumor involvingmedial part ofleft orbit, leftethmoidealsinuses, upperportionoftheleftnasalcavity, part oftheleftfrontal sinus and part ofthe tumor wasalsointracranial- adhering to left lamina fibrosaoftheethmoideal bone in theanteriorfossaoftheskull base. The orbital portionof tumor measured 25x10x34mm, itinvolvedupperobliqueoculomotormuscle and displaced and copressedmedialoculomotormuscle. Theintracranialportionof tumor measured 16x12x10mm. Due to easyaccessiblenasalportiontheinitialprobatorybioptic sample of tumor wastakenfromnasalcavity.According to initial MRI scanthedignosisof ether lymphomaorsarcomawasclinicallyconsidered.Biopticexaminationwasquitepleasantlysurprising and showedmeningiomaofthemeningothelial type, WHO grade 1.Thesurgerywasthenplannedas not extensivelyradicaldue to benignbiologicalnatureofthe tumor and itconcentrated on thereductionoftumorousmass by removingoftheextraorbitalportions. Theattempt to removeintraorbitalportionof tumor wouldprobablyresult in necessity to removetheeye, otherwisehealthy and fullyfunctioning, becauseofinvolvementofoculomotormuscles. So, the orbital portionof tumor was not surgicallyremoved and othermodalitiesofit´s management are stillconsidered. Posibility ofpartialregressionofthe orbital portionisconsidered, and patientisstillunderfollow up. Slightregressionoftheproptosisisobservedaftersurgery, paralyticstrabismwithdiplopia (onlywhenlooking to therightside) persists, as well as normalintraocularfindings.

Pathology: Gross examination: initiallytherewasobtainedonlysmallprobatory sample forthehistologicaldiagnosis, measuring 10x4x3mm. Afterestablishingofbiopticdiagnosissurgicalresectionofthe tumor wasplanned and performed. The specimen fromthisresectionconsistedofmultiplefragmentsfromnasalcavity, ethmoideal

sinus, intracranial part of tumor and adherentsmallfragmentsofthe orbital portion. Theoverallvolumeofresectedtissueswasabout 55x40x20mm. Fragmentsofboneswereinvolved in thematerial.

Lightmicroscopy: the tumor wascompousedofpolygonalcellswithpoorlyopticallydefinedborders andblandmicroscopicalappeareance, withoutatypia, arranged in varyingsizedirregularlobules, withonlyminimalmitoticactivity (up to 1 mitosis per 10 highpowerfields). The tumor filledintertrabecularspacesoftheresectedbones and growedthroughtheerrodedthinosseousbarrier, however, necrosis and agresiveosteolysiswas not noticed. Themicroscopicalappeareanceofthe tumor wasconsistentwithdiagnosisofmenigioma, it´smeningothelial type.
Immunohistochemistrysupportedthisdignosis by typical positivity for EMA and progesteron receptors. Proliferation marker Ki67 was positive in 5-10% oftumorouscells (in theirregulardistribution). Negative results in thetumorouscellswereobtained in theexaminationsfor CD34, ERG, GFAP, S100, cytokeratins AE1/3, CK7,CK5,6, p40, p63, chromogranin, synaptophysin and NSE.
The**finalbiopticdiagnosiswasmeningiomaofthemeningothelial type, WHO grade 1.**

Discussion and Coment: Meningiomaisthe tumor whichsometimesoccurs in the orbital region, most commonlywithorigin in theoptic nerve sheathwhichiscontinuousofmeninges, bothdevelopmentaly and anatomically. In our case the tumor is not connectedwiththeoptic nerve. Optic nerve is not involved in the tumor and theonlyfunctionalproblemsofthelefteyeballisit´sdisplacement and problemswith motilitycaused by oculomotormusclesinvolvement. It seems very likely in our case thattheoriginof tumor isintracranial, in theanteriorfossaabovethe lamina cribrosaoftheethmoideal bone. Whatis very unusual on our case istheextensiveextracranialprogressionof tumor throughtheskull base to thenasoorbital region, despitethefactthat tumor ishistologically WHO grade I, whichmeanslessagresive. Also in thehistological specimen there are not presentnecroses and agresiveosteolysis. Unusuallyprevailingextracranialextensionofthe tumor wasprobablyenhanced by very thin and cribriformshapeofthe lamina cribrosaoftheethmoideal bone, whichwastheosseousstructureunderlyingthe place oftumorousorigin. Whatremainsmatterofquestions, discussions and problemisanother management oftheremaining orbital part ofthe tumor. It was not surgicallyremovedbecauseofinvolvementoftheoculomotormuscles. So, it´ssurgicalremovalwouldprobablynecessitatetheenucleationoftheotherwiseunaffected and fuctioningeyeball and partialexenterationofthe orbit. Such mutilatingsurgerywasabandonedunderthe influence ofotherwisefavourablehistologicaldiagnosis. Therewasdecided to follow up carefullyanotherbehaviorofthe tumor withsome hope thatregressionofthe orbital part of tumor couldbestarted by it´sdebulking and removalofit´sotherparts, eventuallythatthisregressionorat least slowingofit´sprogresscouldbe in futurestimulated by carefullytargeted and carefully dosed actinotherapy.

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